Ophthalmic Rete Mirabile: The First Angiographic Documentation of Embryonic Ophthalmic Collaterals in a Patient with Moyamoya Disease

A Case Report

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Summary

Plexiform vascular networks of the rete mirabile in humans are rare but can be observed in some vertebrates such as cobaye, cat, cow and sheep. Each set of embryonic arteries can be a potential source of these vascular networks which reconstitute the distal flows in segmental agenesis of internal carotid or vertebral arteries especially in patients with PHACE (posterior fossa malformation of the brain, facial hemangiomas, arterial anomalies, cardiac anomalies with coarctation of aorta, and eye abnormalities) syndrome. Ophthalmic rete mirabile has been found in some mammals such as camel, deer and avian species of pigeon, Hawaiian seabirds and deep-water fish of the cod other than human.

We describe the first human case of ophthalmic rete mirabile confirmed on angiography in a patient with moyamoya disease. These findings are the first description in the English literature as far as we know. The clinical implications with moyamoya disease will be briefly discussed according to the embryologic development of cranial arteries.

Case Report

A 42-year-old woman was admitted with recurrent partial seizures. She was born to healthy parents and presented no family history of cerebrovascular diseases. She had suffered from several episodes of brief transient ischemic attacks (TIAs) at 18 years old, and had been treated conservatively in an oriental medicine clinic without anticoagulants. At that time she had not performed cerebral angiograms or CT scan. She had no abnormal findings on physical examination. Blood pressure was within normal range.

On neurological examination and EEG findings, she had partial seizures without any specific epileptic discharges or spikes, representing a seizure focus.

Digital angiography showed complete occlusion of the internal carotid arteries from the region of the carotid fork, with a small number of basal collaterals appearing in classical "puff of smoke" patterns (Figure 1). Injection of the right carotid artery demonstrated the ophthalmic rete mirabile reconstituting the stem of the ophthalmic artery and primitive ventral ophthalmic artery in the medial and inferior olfactory fossa. These collaterals were also connected into basal collaterals of the recurrent artery of Heubner territory.

The ophthalmic rete mirabile was located posterior to the normal retinal blushes on lateral projection and also positioned the most medial part of the inferior olfactory fossa of primitive olfactory artery territory in anteroposterior projection (Figure 1).

Discussion

Plexiform vascular networks of the rete mirabile in humans are rare but can be observed in some vertebrates such as cobaye, cat, cow and sheep. Each set of the embryonic arteries can be a potential source of these vascular networks, so called "rete mirabile", which reconstitutes the distal flows in segmental agenesis of internal carotid artery. The first few human cases of rete mirabile were associated with a hypoplastic cervical and petrous internal carotid artery; a rich anastomotic intradural vascular network at the skull base and a cavernous internal carotid artery that bypassed the absent arterial segments. (Minagi and Newton, 1966; Danziger et Al, 1972) Recently, the rete mirabile has become a relatively well-known vascular network that develops in segmental agenesis of carotid or vertebral arteries especially in patients with PHACE syndrome. (Bhattacharya et al., 2004; Weon et al., 2005) Ophthalmic rete mirabile (ORM) has been described in the literature of some mammals and seabirds other than human (Pettit et al., 1981). This common morphologic configuration of ORM of Hawaiian seabirds suggests that countercurrent heat exchange is the underlying mechanism for cooling the brain, which enhances a bird's tolerance of environmental heat stress during long-distance flight. The common carotid arterial supply to the ORM is derived from a unique intercarotid anastomosis of avian species as birds do not possess circle of Willis collaterals comparable to mammals and human. Therefore, if those morphologic characteristics develop in human they will theoretically explain the embryonic persistence of ORM and consequently it could be responsible for cerebral collateral circulations arising in occlusive cerebrovascular diseases such as moyamoya disease.

Since 1948, when Padget DH who had been a neuroembryologist and neurosurgical illustrator published the first major work on neurodevelopment of the cerebral arterial and venous systems and that contributed to provide a basic anatomical understanding of cerebrovascular disease.

(Kretzer et Al, 2004) Padget had described embryonic developments of the ophthalmic artery with six stages and stage I-II is of utmost importance to explain the findings of ophthalmic rete mirabile. (Padget, 1948) The large primitive dorsal ophthalmic artery (PDOA) takes the main blood supply in 4-5 mm embryo size in stage I development of the embryonic ophthalmic artery. Stage II of 9 mm embryo size shows the vascular plexus or plexiform networks constituted between PDOA and the primitive ventral ophthalmic artery (PVOA). Those embryonic plexiform vascular networks of the primitive ophthalmic artery can be seen in patients with moyamoya disease and representing ophthalmic rete mirabile as persistent embronic vascular annexations between stem of ophthalmic artery and PVOA. (Chung and Weon, 2007)

Moyamoya disease still has an unknown etiology and was originally described by Japanese doctors. (Suzuki and Kodoma, 1983; Suzuki and Takaku 1969) It is characterized by progressive bilateral stenosis or occlusion of the internal carotid artery with the formation of a vascular network, the so-called typical puff of smoke appearances of moyamoya vessels. Classical basal collaterals recruiting leptomeningeal vessels and deep parenchymal vessels of striatum provide blood supply of deep perforators territory and cortical cerebral hemispheres. These collateral vessels are presumed to form during the prenatal period, but definite etiological prenatal occlusive events have not been elucidated. Recently, an etiological hypothesis was suggested (Chung and Weon, 2007) postulating that the pathophysiologic target arterial segment could be located in the proximal middle cerebral artery (MCA) just distal to anterior choroidal artery (AChA) bifurcation during the stage of 14-20 mm embryo size. Those suggestions were based on the angiographic findings considering embryonic development of the cranial arteries in patients with moyamoya disease. Primitive embryonic arteries with remnant anastomosis and annexations were frequently visualized in patients with moyamoya disease especially previously unrecognized ophthalmic collaterals including ophthalmic rete mirabile and PVOA anastomosis with remnant primitive olfactory arteries (Chung and Weon, 2007).

The functional importance of ophthalmic rete mirabile in moyamoya disease is unknown, but these newly discovered anatomic configurations might possibly involve a dynamic role with clinical symptoms emerging as the cerebral ischemia develops in moyamoya disease patients.

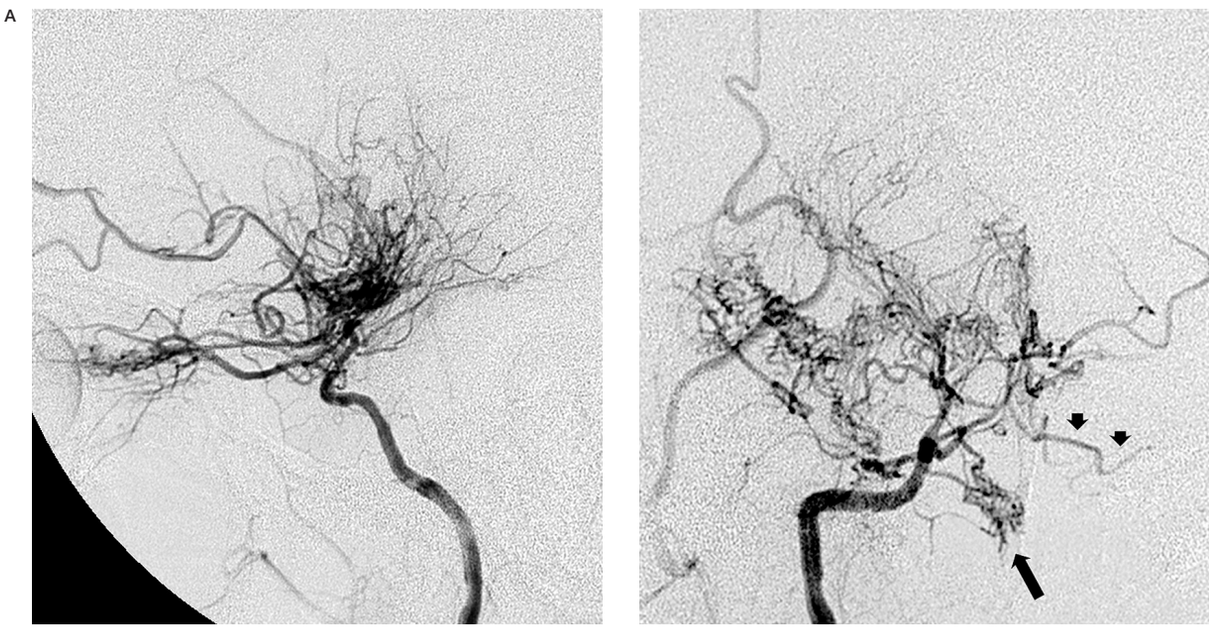

Figure 1 A,B) Lateral and anteroposterior (AP) projection angiograms of moyamoya disease. Injection of right carotid artery shows the ophthalmic rete mirabile located posterior to normal ciliary blushes in lateral projection and medioinferior olfactory fossa in AP projection. Also typical occlusive changes of right supraclinoid internal carotid artery involving carotid fork regions with "puff of smoke" pattern of basal collateral vessels in base of the brain. (A). Injection of the right carotid artery shows the ophthalmic rete mirabile (arrow) located in medioinferior olfactory fossa medial to ophthalmic carotid artery in anteroposterior (AP) projection. Intercarotid anastomosis consisting primitive remnant olfactory arteries (arrowheads) that frequently visible in avian species are visualized in midline frontomedial cerebral hemispheres, which typically courses along the leptomeningeal surfaces (B).

The ophthalmic rete mirabile in our case was typically located in the medial and inferior olfactory fossa. Those findings could be explained by the more medial location of the embryonic orbital cavity and primitive ophthalmic arteries adjoining close proximity with branches of the primitive olfactory artery in stages I-II embryonic developments of the ophthalmic artery. When the ophthalmic rete mirabile persists into postnatal life, the primitive ventral ophthalmic artery arises and connects from it. These findings are based on the embryonic development of primitive ophthalmic arteries in the stages between stage II (9 mm embryo size) and stages III-IV (14-18 mm embryo size). Moreover, anastomosis of the anterior communicating artery in human is completed in the late embryonic period of 24-40 mm embryo size. (Padget, 1948) Absence of the completed anterior communicating artery and A1 segments in moyamoya disease verifies the presence of embryonic remnant annexations and anastomosis between the primitive ophthalmic artery and primitive olfactory artery. Therefore, the leptomeningeal collaterals constituting primitive olfactory arteries are easily recognized in inferomedial surfaces of frontal lobes in moyamoya disease. Comparatively, the common carotid arterial supply to the ophthalmic rete mirabile is derived from a unique intercarotid anastomosis of avian species as birds do not possess a circle of Willis collaterals. This finding is of interest because the ophthalmic rete mirabile may represent an anatomic homologue comparable between avian physiologic normal ophthalmic vessels and human embryonic remnant ophthalmic collaterals especially in cases of moyamoya disease.

The angiographic findings of ophthalmic rete mirabile were described in two cases as undetermined ophthalmic collaterals by Korean neurosurgeons (Lee et Al, 1975). In that time era, the definition of moyamoya disease was ongoing and its variable angiographic manifestations had been largely unrecognized. We also possibly identify the same morphologies of ORM in another paper (Silbergleit et al., 2000) but it has never been documented precisely.

There are no reports in the English literature on ophthalmic rete mirabile to our knowledge, which are frequently visualized in patients with moyamoya disease. (Chung and Weon, 2007)

Conclusions

In conclusion, we describe the first human case of ophthalmic rete mirabile presenting ophthalmic collaterals between the stem of the ophthalmic artery and primitive ventral ophthalmic artery in a moyamoya disease patient. The ophthalmic rete mirabile could be visualized as embryonic remnant anastomosis and annexations at an earlier time of the development of cranial ophthalmic arteries thereby explaining the prenatal occlusive events of undetermined cause such as moyamoya disease.

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